ASTHENIC BULBAR PALSY, WITH REPORT OF A CASE.*

By John Punton, M.D.,

KANSAS CITY, MO.

A few months ago the case I am about to report was referred to me by Dr. L——, of Columbus, Kansas, for diagnosis. In the course of my examination I discovered that the history and symptomatology were unlike that of anything in my previous experience, although my reading suggested to me what I think the case really is.

In order to confirm my suspicions I called to my aid my colleague Dr. Burnett, to whose co-operation I am mainly indebted for this detailed report. It is not my desire, however, in reporting the case to claim anything new or original, but simply to call attention to, and to put on record, one more case of this relatively rare form of nervous disease of which we have considerable yet to learn from a pathologic standpoint.

It is only within the last two decades that this affection has actually been impressed upon us as a distinct neurologic entity deserving a differentiation from that of Duchenne's disease, or true bulbar palsy, although Wilks[1] in 1870 presented a clinical résumé plus a post-mortem study that was followed a little later by an analytical study of three cases by Erb, whose combined efforts to gain special recogniti n of the disease fell flat. Only after Oppenheim, a decade later presented his non-confirmatory microscopical findings, showing the symptom-complex to be without a distinct anatomic change, was the stimulation sufficient to produce the series of papers that followed, illy describing the condition and leading to confusion in the production of a wide-spread nomenclature. Really, not until 1892[2] did the disease receive special recognition, this being prompted by Hoppe's[3] paper. Dr. Collins in 1896 speaks of a dozen cases on record in which post-mortem examinations have been carefully made. While our patient is still living, and therefore the case offers no pathological evidence, it is clear that the clinical analysis is sufficient to demonstrate wherein the case differs from a chronically progressive degenerative bulbar paralysis, characterized by fibrillary twitchings, atrophy, and paralysis; and that it is almost a typical picture of those cases of asthenic bulbar palsy as verified by necropsies. The case in point is as follows:

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*Read at the twenty-fifth annual meeting of the American Neurological Association, June 14 and 15, 1899.

[1] Wilks, Guy's Hospital Reports, Vol. XXII.

[2] Collins, International Medical Magazine, April, 1896.

[3] Hoppe, Berliner klin. Wochenschrift, 1892, p. 332.

Family History.—Grandparents, dead; no knowledge of the cause of death. The mother of the patient is asthmatic; the father is a strong healthy man.

Personal History.—The patient, Mrs. B., is twenty-five years old, married happily, and the mother of one child three years old. Prior to marriage she suffered, at intervals, from attacks of so-called rheumatism, characterized by pain in the joints without swelling; otherwise she was an average healthy girl. Her life has been devoid of any special hardships or mental depression.

The symptoms of her present ill health date back about three years. Shortly prior to this time she gave birth to her first and only child. The parturient act was prolonged and difficult. A midwife officiated, leaving the usual lacerations as souvenirs of her skill. Operation twelve months later for lacerated cervix was done successfully, while she was being treated at Hot Springs for her supposed rheumatism. She believes that she has not been well since her delivery. The first manifestation of the condition now complained of was an affection of the eyes, coming on at intervals, when she would see double, and her vision would become more or less dim—a manifest weakness of her ocular muscles. Ptosis of left eye supervened, being also worse at intervals. Her general health began to fail, and amyosthenia was marked. She became profoundly weak on the slightest exertion, the extremities seemed like heavy burdens; mastication, which at first appeared normal, gradually became fatiguing, swallowing became difficult and imperfect, and articulation also became impaired. The voice changed in pitch, tone and quality, being distinctly nasal in character. The tongue became less dexterous and was the seat of "numbness," as she explains it. The prehensile acts performed by it during mastication were limited, imperfect and fatiguing. The food could neither be removed readily from the buccal grasp, nor be conveyed posteriorily to the constrictors, which in turn were functionally derelict, and deglutition, especially of solids, was markedly unsatisfactory. While abnormal weariness, prostration and rapid heart action are induced by slight efforts of bodily occupation, she has no morbid fears, apprehensions, dreads or perverted conceptions; neither is she moody, despondent or given to morbid mentality. In two years from the onset of the disease she had ptosis of the right eye and weakness extending over the facial musculature. She says the movements of her lips gradually became limited until she could no longer co-apt them as in the act of whistling. The strabismus developed in both eyes, and the movements of the eyes became limited in all directions. Since this defect of the eyes there gradually developed a continuous lachrymation. She says she has decreased in weight from 153 to 106 pounds.

Examination.—On close examination we found her eyeballs fixed, there being no movement in any direction. They were also slightly protrusive. Excessive lachrymation was constant during the waking hours. The pupils reacted to light and in accommodation, and ptosis of left eye was marked; slightly so in the right. The diplopia, formerly so annoying, is now only present when certain lateral head movements are made since ophthalmoplegia has supervened.

Face.—The whole facial expression suggests a putty-like blankness. The muscular tonicity is lost, so that a state of profound paresis on the right side exists with a less degree of paresis on the left. The lips cannot be co-apted sufficiently to produce labial sounds, neither can the patient whistle. Attempts to show the teeth or to produce laughing expression distort the face slightly to the left. The masseters are about equally paretic and it is impossible for the superior and inferior maxillæ to be approximated with any degree of force. The chewing of bulky and solid substances cannot be done for want of power. There is no atrophy, tremor or fibrillary twitchings of the muscles involved.

Organs of Speech and Deglutition.—The tongue is thick and flabby, without indentures, atrophy or tremor, and is utterly devoid of any lateral movements. It can only be protruded to the inner margin of the lips. The palate has lost its symmetrically arched outline, and hangs low, with the uvula directly on the base of the tongue, and does not rise to close off the posterior nares in attempts to say "Oh," etc. The voice is exceedingly thick, and all sounds are indistinct, and finished off with the cleft-palate nasal twang. Her attempts to answer the necessary questions in the examination cause her much fatigue. The constrictor muscles of the throat are quite paretic, and deglutition of solids is difficult, while liquids regurgitate through the nose. A careful examination of the organs of voice by Dr. Hal Foster revealed paralysis of the left vocal cord.

Trunk and Extremities.—She is thin, as would be expected when remembering she is but a medium-sized woman, having lost 47 pounds in weight. There is no atrophy of shoulder, arm, forearm, hands or trunk muscles, neither are there any fibrillary twitchings to be seen anywhere on close and repeated examinations. The power in all the extremities seems normal to the usual tests for one in her physical health, and there are no sensory symptoms present.

Reflexes.—The superficial reflexes are normal, whilst the deep reflexes (knee-jerks) are slightly exaggerated. No ankle-clonus.

The electric irritability of muscles is normal, and there is no evidence of disease of the heart or lungs. Urinary analysis proved negative. The pulse was 90, and regular. Menses and bowels were also regular. Appetite fair, notwithstanding the great loss in weight.

It would seem that while this patient was subject to remissions and exacerbations since the advent of her affliction, these variations have not been so markedly manifest as in the average case. When she did enjoy an apparent remission it was more of an arrest of the progress of the disease for a time only, to be broken by a sudden onward march to the decline. There appears to have been no special epochs when there was a cessation of her symptoms, with pronounced steps toward recovery, as was noted in Collins' report. It is true her amyosthenic manifestations were aggravated more at times than at others, but these were really brought on by slight efforts causing fatigue, and were largely corrected by removing the fatigue. The knee-jerks in this case while absent on the first test were later exaggerated, but grew less responsive on repeated tests, however, until only a minimum reflex act could be obtained. They were also restored to former activity by a period of rest. Any attempt to tax any organ or organs was soon followed by distressing fatigue and general nervousness. Severe attacks of tachycardia with prostration and weakness were sure to follow over-expenditure of her limited strength; but that there was actual improvement at any of the seeming periods of better health over and above arrested progress, there is no evidence.

A symptom attracting attention was the profuse lachrymation, a symptom common in true bulbar palsy, but usually attended by more or less emotion, which was wanting in this case. She was perfectly clear and responsive mentally, and remarkable for her buoyancy and composure. It must be remembered that complete ophthalmoplegia existed, and while ptosis was marked, there was a protrusion, seemingly of a naturally large eyeball, which remained unprotected from the particles of atmospheric dust ordinarily whipped away by the physiologic activity of the eye lashes. Exact observations on the working of the vocal cords are largely wanting in the literature on this subject; the nearest expression is the supposition of a paretic condition of the vocal cords, such as mentioned in Hoppe's case. Of course, paralysis of the palate, tonguê and lips is disastrous to perfect externalization of speech, but the case presents the additional defect of paralysis of the left vocal cord. To be positive of this the competent counsel of Dr. Hal Foster was asked. In attempts at phonation the left vocal cord refused to functionate, was relaxed, passive, and did not rise to meet its fellow of the opposite side in the median line, and presented a pale anemic appearance. The right vocal cord, on the other hand, presented no abnormal appearance, and in functionating passed well across the median line in an attempt to approximate its derelict companion.

At a glance it will be seen that the symptom-complex making up the clinical picture is purely a motor defect. It is a typical denotation of suspended motorial function. As already mentioned, no evidence of superficial or deep sensory defects were manifest. The complete ophthalmoplegia externa (there being normal response of the pupil to light reflex and in accommodation) points to involvement of the third and sixth nuclei; bilateral weakness of the muscles of mastication sufficient to prevent the chewing of solid substances implicates the motor branch of the fifth; the facial palsy on one side and distinct paresis on the other, giving unilateral facial deflection on muscle functionation, and the blank non-expressive appearance in a state of quietude, is due to suspended innervation from the seventh nuclei; and a paralytic state sufficient to prevent phonation, vocalization, articulation and deglutition points to the 9th and 10th nerves, whose motor fibers arise in common from the nucleus ambiguus, which is a continuation of the motor cell-column of the anterior horn, while the 12th nerve is responsible for the lingual disability.

Diagnosis.—In the diagnosis of asthenic bulbar paralysis we are called upon to differentiate more particularly from pseudo-bulbar paralysis and true degenerative bulbar paralysis. From the former, as Dr. Collins says, "it must always remain a difficult question, as in both there is absence of atrophy in the paretic muscles and in both the symptom-complex may be only atypically that of true bulbar paralysis."

Mills[4] calls attention to the fact that in the majority of cases of pseudo-bulbar paralysis reported, the patients were aged and their vessels highly atheromatous. He also states that "one of the most important points of distinction between true bulbar paralysis and pseudo-bulbar paralysis is the condition of the tongue, which in the former becomes markedly atrophied and is the seat of fibrillary contractions from which in the latter it is free."

While it is true that a striking feature in our case was the absence of atrophy in any of the affected muscles, yet there was evidence of paresis of every motor cranial nerve beginning with the third and ending with the twelfth. The face, shoulders and limbs were remarkably well rounded, considering that the patient had lost 47 pounds in weight. The muscles supplied by the facial, motor fifth, and third nerves were, however, markedly affected, but the effect of the paresis was somewhat transient in character, the patient partially recovering the normal use of these muscles between the paroxysms, which according to Mills is the rule in the asthenic type.

In genuine bulbar paralysis the hypoglossus, vagus and glossopharyngeus are more seriously and continuously affected. No positive evidence of fibrillary twitchings could be elicited as being present, or as having been present. The reflexes were retained with the knee-jerks plus. The electrical response was normal. The psychical faculties were in no wise affected. The ciliary muscles were not involved, and the vision was normal. There was no dribbling of saliva. The tendency to fatigue and prostration was even more manifest on the slightest exertion, especially in muscles supplied by the motor cranial nerves, while to some degree the whole organism suffered. There were no disorders of common sensibility or special senses. Thus it would seem that the case is not one of glosso-labio-laryngo-pharyngeal paralysis, but is the clinical picture of a genuine case of asthenic bulbar paralysis.

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[4] Mills, Text-book on Nervous Diseases.

DISCUSSION.

Dr. Joseph Collins thought that asthenic bulbar paralysis was a sufficiently rare disease to warrant the recording of a new case. The case that Dr. Sinkler reported seemed to parallel the one published by Dr. Collins, except in one or two particulars. Dr. Collins did not believe that the exhaustibility of the muscles by the faradic current, as pointed out by Jolly, would be corroborated by future investigations. In Dr. Collins' case the muscles were exhaustible on certain days, but on other days no more exhaustible by the faradic than by the galvanic current, though more so than in the normal individual. Then the knee-jerks were continually present in Dr. Sinkler's case, but in Dr. Collins' case they were present when the patient was comparatively well; absent when she had bad days, and exhaustible at all times; *i. e.*, they became less vigorous under repeated blows and after the seventh, eighth or ninth tap were entirely exhausted.

Dr. Collins was not inclined to look upon the disease as one of the motor nerves primarily. It seemed to him that there is very little in the clinical manifestations to warrant us in taking the disease out of the domain of the sympathetic nervous system. True, its phenomena are manifested largely through the motor system, but its most important symptomatology is due to trouble in the sympathetic system. That had been firmly impressed upon Dr. Collins in the observation of his own case. He sometimes found his patient in a state of collapse, with ballooned abdomen, blue lips and blue finger tips, cold extremities and feeble pulse; and if it were not for the subcutaneous injection of large amounts of normal salt solution and the use of diffusible stimulants, he was sure she would have died just as patients do from surgical shock.

The most plausible theory of the pathogenesis of the disease, it seemed to him, was that in all probability there is some poison or infectious material generated within the system, continuously operative upon the vegetative portion of the body. This toxic material manifests itself periodically, and whenever the vitality is sufficiently low to allow the poison to get the upper hand, the phenomena of relapse appear. What the source of such a poison is, we can only conjecture. The point brought out by Dr. Sinkler, that there was a weakness of the nervous system manifesting itself in other members of the family, would only go to show that this disability may be in part inherited as well as acquired.

The remarks that Dr. Sinkler made concerning the necessity for differentiating the disease from hysteria seemed to Dr. Collins to be very well taken. The case reported by Dr.

Collins had been treated by specialists and general practitioners as one of hysteria.

A symptom that had not been mentioned in the papers read was one in relation to the menstruation. Dr. Collins' patient was always worse at the menstrual periods. She had great menorrhagia and on two occasions he had to pack the uterus to stop the hemorrhage.

Another very remarkable symptom or group of symptoms in his patient occurred in the shape of an acute psychosis, taking the form of confusional insanity. Any attempt to explain the occurrence of this must be in line with the statement that the poison which is in the system accumulates and at the time of the paroxysm exercises its effect upon the cortical cells as well as upon the other cells of the central nervous system.

Dr. W. G. Spiller referred to the fact that Dr. Sinkler had laid especial stress on a symptom that is very important in asthenic bulbar paralysis—the symptom of fatigue. In Dr. Punton's case restoration of function after rest was not nearly so prominent and Dr. Spiller was not at all certain that Dr. Punton's case was not one of pseudo-bulbar paralysis or possibly one of "bulbar paralysis without anatomical findings." The case, however, might be one of asthenic bulbar paralysis. The bulbar diseases still need much study.

Dr. Spiller said that a necropsy had been obtained in a case of asthenic bulbar paralysis reported by Widal and Marinesco, and changes in the nerve cells had been observed.

Dr. H. M. Thomas said he had had a case under observation for two years that seemed to him to be one of asthenic bulbar paralysis in the early stages. No permanent paralysis of the motor nerves of the eye or of the muscles of the face was present. The patient was in pretty good condition on getting up in the morning. She could speak fairly well for about half an hour, and then her voice gave out. When she began to eat her breakfast, she ate well enough until she had masticated for a few minutes, and then it became progressively harder for her to chew. If she was required to look up more than four or five times in succession, ptosis became evident and weakness of the superior rectus of each eye developed. Unfortunately the patient lived in another city and he had not been able to see her for a year, but had heard that she had periods of improvement and relapse.

Dr. Thomas had also had under observation another case that puzzled him a great deal as to diagnosis. The patient was a young girl who had the symptom of fatigue well marked, more particularly in the muscles of the extremities than in the muscles supplied by the cerebral nerves. She had never had a complete remission, but was always better in the morning before she took any exercise, and got progressively worse towards evening. Dr. Thomas believed that the case was not one of muscular dystrophy.

Dr. Sinkler did not wish to go into a discussion of the pathology of the disease, as it was too vague and uncertain. He only wanted to refer to two symptoms in his case that he failed to emphasize sufficiently, namely, the absence of any muscular tremor even in the tongue, and the absence of atrophy.

Dr. Punton said that the question of syphilis was very carefully considered in his case and also the question of pseudobulbar paralysis, and they had come to the conclusion that the case did not belong to either class, and that it was one of asthenic bulbar paralysis.

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173. A PROPOS DE LA CHORÉE VARIABLE DE BRISSAUD. TROIS OBSERVATIONS DE CHORÉE (On Brissaud's Polymorphous Chorea. Three Observations on Chorea). A. Couvelaire et O. Crouzon (Revue neurologique, 7, 1899, p. 399).

Brissaud in 1896 described a new type of chorea, differing from that of Sydenham and of Huntington, which he called the "chorée variable des dégénérés." This new type, which may be termed "polymorphous chorea of degenerates," is characterized by the multiplicity and variety of its movements, by its remissions and variability in time, and also by the presence of various stigmata of degeneration in the subject affected. The authors present three cases from Marie's clinic, at the Bicêtre, illustrative of the condition. The first occurred in an epileptic. Her movements were bizarre and at times extremely violent, not permitting her to hold any articles in her hand. She had attacks of petit mal. The chorea was cured. The second case is termed a chronic non-progressive chorea. In this case the movements were greatly accentuated, though at first sight no marked abnormality could be detected. On close scrutiny, however, quick movements of the hands, lips, fingers, shoulders, eyelids, could be detected. At times the patient could not sit quiet. There were no degenerative stigmata in this case. The third case is one of irregular Sydenham's chorea. The second case more closely corresponds to Brissaud's type, though it lacked the stigmata. JELLIFFE.